

ARTICLE II.

A CONTRIBUTION TO JACKSONIAN EPILEPSY AND THE SITUATION OF THE LEG CENTRE.¹ By WILLIAM OSLER, M.D., F.R.C.P. LOND., Professor of Clinical Medicine in the University of Pennsylvania.

THE case here recorded illustrates the following points : Epileptiform seizures from a very limited lesion ; the situation of the leg centre, and certain features in the clinical history of the disease.

The present doctrine of cerebral localization may be said to have had its origin in the study of the effects of very limited cortical lesions, and the labors of Fritsch, Hitzig, Ferrier, and others have removed the subject from the region of speculation to the solid ground of experimental science. Still, as far as man is concerned, while admitting the great and corroborative value of observations upon dogs and monkeys, the careful study of pathological cases offers the only means whereby positive knowledge can be attained. Year by year in the past decade evidence of this nature has been accumulating, and more important results may be expected as the records become more exact and scientific. Fully twenty years ago Dr. Hughlings-Jackson, studying cases of unilateral convulsions or spasmodic seizures limited to one member, found that they were often associated with localized spots of disease on the surface of the brain, and he suggested, in explanation of such cases, that the lesion was of the nature of an irritant to the cells of the gray-cortex, which discharged themselves, so to speak, in an irregular and explosive manner, causing a convulsion or spasmodic action of the muscles over which they normally presided. As the seizures began either in the arm, leg, or face, it was reasonable to conclude that the portion of the cortex affected was different in each instance,—*i. e.*, there were actually centres—motor in character—which when irritated in this way caused the convulsive attacks.

When experiments on animals demonstrated that the gray matter was irritable, and that stimulation of limited areas was followed by contraction of definite groups of muscles, Dr. Jackson's suggestion of motor centres was seen in its true light. Ferrier's observations on monkeys enabled him to indicate approximately the homologous motor centres in the human brain, and an extraordinary impetus was thereby given to the study of cerebral cases bearing upon localization. The result of the ten or twelve years' work enables us to speak with some degree of positiveness of the functions of certain regions of the brain. Thus the motor area has been ascertained to be in the mid-region embracing the convolutions on either side of the fissure of Rolando. Irritative lesions of these parts issue in convulsions more or less limited, destructive lesions cause paralysis, local or generalized

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according to the extent of the disease. The other areas of the cortex cerebri are silent, *quoad* motor effects when stimulated, and when destroyed do not necessarily induce paralysis. With regard to further specializing of centres in the motor region, as far as man is concerned, the analysis of cases would appear to place the leg centre in the upper part of the central convolutions, particularly the part extending to the median surface—the paracentral lobule; the arm and hand centre in the mid-region of the central gyri, and the centres for the face and tongue at the lower end—a disposition in each instance coinciding more or less closely with the conclusions arrived at by Ferrier from his observations on monkeys.

Dividing cerebral symptoms into those accompanied with loss of function—negative, and those characterized by excess of function—positive, the cases of cortical epilepsy may be taken as examples of the latter group. In Dr. Jackson's phraseology, the proximate cause of the paroxysm is an abnormally highly unstable condition of the cells of the gray matter, resulting in a sudden discharge. "Healthy movement implies a liberation of energy or nervous discharge initially by cerebral cells, at any rate if the movement be a voluntary one. A convulsion, that is to say, a sudden, excessive, rapid, and temporary development of movements—many movements 'run up' into spasm implies of necessity a corresponding, sudden, etc., discharge." In a local spasm only a few cells are in this highly unstable condition; in severe seizures the sudden and excessive discharge of the highly unstable cells overcomes, it is supposed, the resistance of healthy cells in physiological connection with those highly unstable.

These preliminary remarks will enable the history of the case to be more satisfactorily followed, and I may state too, the main points of difference between these epileptiform seizures and true epilepsy; the slow onset, local in character, beginning in, or in mild attacks confined to, one limb or a single group of muscles; the gradual extension until the side is involved, or, in severe attacks the entire body; loss of consciousness late, not early and sudden as in true epilepsy, and lastly, the muscular contractions are clonic, rarely or never tonic.

On November 8, 1883, I received from Dr. — the brain of his daughter for examination, and with it the following history:—

E. L. M., aged 15 years 9 mo. When sixteen months old fell on her head from a table and appeared to be very much hurt, as she cried violently for a long time after. She appeared to be quite well for about five months, when the left hand was noticed to close firmly, and it seemed to pain her a little from the firmness of the contraction. This continued to increase in severity and frequency for three months, when the left leg became similarly affected, and in two months more she was confined to bed, and the paroxysms had become general all over the body, the mouth being generally fixed open during a spasm.

These spasms lasted in this violent form for about two months, she having as many as eight or ten in an hour. There never was at any

time any loss of consciousness. This makes about seven months altogether. Then suddenly the whole trouble ceased, and she was perfectly well and ran about as healthy a specimen of a child as could be seen.

She remained quite free from spasms for one year, when they returned in the same way, and ran much the same course for six or seven months, and then she recovered perfectly again for about the same length of time, and this went on till she was about eight years of age, or about six years after the first illness, when the left leg began to show signs of weakness and gradually the foot turned in, but she still ran about. •

To give an idea of the kind of spasms she had about that time, I will describe one:—

Suppose her at the dinner table, she would suddenly say, “Oh, I am going to have a spasm.” (She knew this by the contraction of the left hand.) She would then jump up and go to the sofa, get a cushion, lay it down on the floor, then lie down with her head on the pillow, and jerk away in a spasm for half a minute or a minute laughing or talking all through it, and never losing consciousness. She would then get up, replace the cushion, and come back to the table and finish her dinner.

After each interval, of many months, the seizures were more severe; and shortly after she attained her eleventh year, there was a return of the illness, which never ceased for nearly four years, and during six weeks of that time she lay unconscious, and had from fifty to eighty spasms during each twenty-four hours; but as soon as they became less frequent, she became perfectly conscious, and was able to sit up in bed or an invalid chair and read or do a little fancy work, although the left hand was very feeble, and the joints of the fingers would bend nearly as far backwards as they would forwards; this condition of the joints being the result of the position assumed by the fingers during the seizures.

Last Christmas, when she was nearly fifteen years of age, the spasms suddenly ceased, and she was for ten months without them, and during that time she became fat and rosy.

During all these years she was a remarkably intelligent child, and even very much above the average, for without any education of any consequence she was far beyond those of her age. Her memory was something remarkable.

There were no signs of disease on the body, excepting that the skin of the legs became very rough after the seizures commenced and disappeared after they ceased.

During the last two years the toes of first the right and then the left foot assumed a brownish-yellow appearance, which no amount of washing would remove, and latterly the skin became thickened, and small sections of this dirty brown epithelium peeled off and soon re-formed.

There was very little, if any, wasting of the limbs of the left side, but the foot was flexed inwards at a right angle to the leg, at last, and firmly flexed in that position.

Just a week before death, the spasms returned with great violence and increasing frequency, till they became almost continuous, and for two days there was complete unconsciousness or coma. Three hours before death the spasms ceased, and she died very quietly, as I suppose from congestion of the brain, as the conjunctiva were very much injected and the temperature very high. The post-mortem (so I was told) revealed a very much congested condition of the vessels of the brain.

Just a week before she died she told the nurse to be sure to tell me to

have a post-mortem, as she knew her case was a peculiar one, and that it might be of benefit to some one else, and to the medical profession in particular.

In reply to questions, the doctor supplied the following additional information: "The spasms always began in the left hand and *never* in the leg. For about two months at the beginning of the illness the hand just closed firmly for a few seconds, and there was no twitching, but after the expiration of the two months it always twitched from the onset of the spasm. Frequently she could be seen standing with the hand closed and jerking before the leg became affected, and she had to lie down. The spasms were never confined to the left leg. When the leg did become involved the twitching began in the toes and ran up the limb. At the first the arm alone was affected. When the spasms became unilateral, the face would twitch and the eyes roll to the convulsed side. The left arm though feeble was not *stiff*, and in the same useless state as the leg.

The clinical history may be summarized as follows: Jacksonian epilepsy lasting over fourteen years; the convulsions beginning in the left hand, at first monobrachial, then extending to the leg, afterwards becoming unilateral, and finally general, at first without loss of consciousness. For the first nine years of the illness, remarkable intermissions lasting for six or seven months, once an entire year. Six years after the onset the left leg got weak and stiff. For four years, the tenth, eleventh, twelfth, and thirteenth of the illness, the seizures frequent, during this period, six weeks' unconsciousness in which the spasms were very frequent, fifty to eighty in the day. Ten months prior to final attacks freedom from convulsions. Intellectual faculties unimpaired.

Brain examined on Nov. 9th; organ large and well formed; dura natural; hemispheres symmetrical; no special cloudiness of arachnoid; Pacchionian granulations small; large and small vessels of pia mater enlarged, and gave a very congested appearance to the surface; no adhesions of the membrane; no spots of opacity or thickening; the pia mater stripped off exposed natural looking convolutions of a deep pink-gray color; motor convolutions looked symmetrical, no puckering or depression; vessels at base healthy; right crus badly torn. The cord was cut just at junction with medulla, in the lateral aspect of which there is also a laceration; the organ was sliced after the Pitres method. *Pre-frontal* and *pediculo-frontal* sections normal. A section passing 3 centimetres in front of the fissure of Rolando shows nothing abnormal. In making the *frontal* section the knife met with increased resistance on the right side, and the section which passed through the ascending frontal convolution, exactly 2 cm. in front of the fissure of Rolando, exposed a firm fibrous mass occupying the upper part of this convolution in the superior fasciculus of white fibres. It measured 14 mm. in width by 15 mm. in vertical length, was 8 mm. from the surface towards the longitudinal fissure, 10 mm. from the top of the convolution at the margin of the long fissure, and 15 mm. from the external surface. It ran up to the gray matter, but did not appear to involve it except towards the median surface.

In a section 7 or 8 mm. behind the *frontal* the mass was still visible as a small round puckered area, situated just at the edge of the gray matter at the bottom of a sulcus passing into the asc. frontal from the fissure of Rolando, about 15 mm. from the longitudinal fissure. It extended to within 4 or 5 mm. of the fissure of Rolando. Thus the entire mass was within the upper end of the asc. frontal gyrus, having an antero-posterior extent of about 17 mm., and a vertical diameter of 15 or 16 mm., almost entirely within the white substance, but bordering on the gray matter at several places.

Unfortunately the torn state of the crus and medulla made it impossible to trace any descending sclerosis in these parts. Histologically the growth presented the characters of a firm glioma, consisting of 1st, and chiefly, a dense felt-work of fibres, in places coarse and devoid of cell elements; 2d, cells of various sizes, branched and fusiform, the processes of which could be directly traced in connection with the fibres. Towards the peripheral part of the growth the cells were more abundant; 3d, bloodvessels pretty numerous and large considering the amount of fibrous tissue in the mass. The growth shaded into the contiguous tissue in a very characteristic way, and towards the gray matter there was no sharply defined border, although in the microscopic sections it was easy to see where the normal tissue began, and there was a zone in which there were scattered a number of deeply stained small cells like leucocytes. In most of the sections the ganglion cells of the contiguous gray matter looked normal and their nuclei took the logwood dye as usual. On the side of the convolution towards the fissure of Rolando the growth directly involved the gray cortex. A study of the sections did not appear to bear out Klebs's view that the ganglion cells participate in the growth.

The case is unusual in the limitation of the lesion to one convolution and to its fasciculus of white matter, scarcely involving the gray substance which is commonly affected in cortical epilepsy. The accurate localization and the remarkable absence of tissue changes in the immediate vicinity give the case the nature of an exact physiological experiment. It is the rule almost for lesions causing epileptiform convulsions to involve the cortex, such as meningeal thickening and growths, exostoses, gliomas, and other tumors of the surface. They need not, however, directly affect the motor zone, but may be in the vicinity, near enough to excite irritation of the centres. Charcot lays down the following rule for guidance in this matter: When in the intervals of the attacks the patient has not any form of permanent paralysis, the disease causing the convulsions is in the non-motor zone, but when, on the contrary, the patient is paralyzed in the intervals, either monoplegic or paraplegic, we may conclude that there is a destructive lesion of the motor area, more or less limited. For example, a lesion at the base of the second frontal convolution might irritate the contiguous motor cells of the arm centre in the ascending frontal and produce epileptiform seizures without any permanent paralysis; or, if

in the central part of the motor convolutions, might produce irritative effects in the leg and face centres above and below it, while at the same time there was paralysis of the arm from destruction of its centre. In fact from cortical lesions in this region we may have the epileptiform seizures without the paralysis, or there may be paralysis with the seizures, or in some cases limited paralysis without convulsions. In the present instance there was, with a limited lesion of the motor area, permanent paralysis with contracture of one extremity and epileptiform convulsions.

In this class of seizures the spasms may begin in the hand, the face, or the foot, and, according to Jackson, this is the order of frequency, and, as a rule, the attacks begin always in the same place. They may be confined to the one region—monospasm, or may gradually extend until one half of the body is involved—hemispasm. Facial and brachial monospasm are more common than crural. The attacks may be limited at first to a group of muscles in an extremity, or to the entire limb. Thus, in the case of the patient with this disease, which I showed at the society some months ago, there was brachial monospasm, and in the one under consideration, the doctor states that the child might be seen standing while the arm was convulsed.

The order of spreading is important; it is usually up a limb, but it may be in the opposite direction, and in the event of the monospasm extending it is more common for the face to be involved with the arm, or *vice versa*, and the leg with the arm, than the leg with the face. Here from what can be gathered the order of march of the spasm was up the arm, then the leg became affected, and afterwards the face. This is unusual; it is more common for the leg to be affected last. Complete details, however, of the precise sequence of the spasms are wanting. Evidently at first there was brachial monospasm, then extension to the leg, and later hemispasm with rolling of the eyes and affection of the face muscles. Within six months from the origin of the trouble the seizures had become general, but the doctor says there was up to this time no loss of consciousness, such as subsequently took place.

The extension of the convulsions to the other side is explained in one of two ways; either through the direct pyramidal fasciculi with which each side of the brain is connected in a greater or less degree with the same side of the body, or more probably, on Broadbent's theory, that it is owing to "active conditions of the decussating fibres putting in action the associated nuclei of both sides of the cord, and then the bilaterally acting muscles of both sides of the body." The discharge of the nerve cells of the cortex cerebri excites the motor nuclei of the cord, and the violent impulses pass from the spiral ganglia to the muscles. Now it is easy to conceive that when the discharges are excessive and violent, the ganglia of the other side of the cord may be excited through the commissural fibres which unite the nerve cells of the anterior horns.

The long duration, fourteen years, of a glioma, is not without parallel. Dr. Jackson has recorded two cases, in one of which the fits lasted ten, and in the other twelve years. Cerebral gliomata are benign growths, which grow slowly and never produce metastases.

The other feature of interest in this case is the light it throws on the situation of the leg centre. Ferrier placed this in monkeys at the gyri at the upper end of the fissure of Rolando, and the result of pathological investigations in man point to the same situation. Cases of uncomplicated crural monospasm, or monoplegia are not common, but in the observations analyzed and collected by Ferrier and by Charcot and Pitres, the lesion was in each instance in the upper part of the central gyri, or in their extension on the median surface. When this part is simply irritated, there may be spasms beginning in, or limited to, the foot and leg; when the seat of a destructive lesion there is crural monoplegia. In their latest work,¹ MM. Charcot and Pitres bring forward additional evidence in support of this view. In the case here recorded, the fibrous mass was situated entirely within the anterior part of the paracental lobule, limited in extent, confined chiefly to the medullary fibres of the superior frontal fasciculus, and only touched the gray matter in places. A point to be referred to is the absence of the paralysis of the leg for the first six years—for if the convulsions and monoplegia were caused by the same legion, how explain the late onset of the latter? From the fibroid state of the tumor, it might reasonably be inferred that it was originally larger, and had shrunk, but the absence of puckering on the surface, and the way in which the margins merged with the contiguous parts, make it probable that the growth was always small—so small, in fact, that at one period of its development it may have caused sufficient irritation to induce the convulsions, and yet at the same time not involved the special fasciculi of white fibres to the extent of producing weakness of the leg or monoplegia.

In the clinical history, the duration, fourteen years, is the most remarkable feature; it is rare for cases of cortical epilepsy to run such a prolonged course. The irregularity of the seizures, the long intervals and attacks of coma, which characterize so large a proportion of these cases, are phenomena not less difficult of explanation here where a lesion is present, than in cases of ordinary epilepsy in which coarse alterations are not usually met with.

¹ *Revue de Médecine*, Octobre, 1883.